Postoperative Henoch-Schönlein Purpura

Joshua A. Gross, MD; Brittany E. Grady, DO; Katya Sanchez, MD; David J. Gross, MD

PRACTICE **POINTS**

- Henoch-Schönlein purpura is a multidisciplinary problem.
- Henoch-Schönlein purpura is an IgA-mediated disorder that is more common in children and has a more severe course in adults.

To the Editor:

A 57-year-old man with a history of type 2 diabetes mellitus and hypertension was hospitalized for heart disease resulting in an aortic valve replacement and multiple-vessel bypass grafting. He experienced a stormy septic postoperative course during which he developed numerous palpable purplish plaques (Figure 1). The lesions were bilateral and more heavily involved the lower legs and buttocks. The head and neck remained free of skin lesions. Additionally, the patient reported a bilateral burning sensation from the knees to the feet.

Punch biopsies of lesions from the right upper arm were obtained. Hematoxylin and eosin staining revealed neutrophilic-predominant small vessel vasculitis (Figure 2A) with the upper dermal location more heavily involved, as demonstrated by involvement of a superficial vascular plexus (Figures 2B and 2C) that was consistent with Henoch-Schönlein purpura (HSP). The diagnosis later was confirmed with immunofluorescence. Direct immunofluorescence revealed granular IgA deposition around the superficial vascular plexus (Figure 3). No IgG, IgM, C3, C5b-9 complement complex, or fibrinogen deposition was seen. Additionally, periodic acid–Schiff staining failed to show microorganisms, thrombi, or intravascular hyaline material.

At our initial consultation, we observed an ill-appearing afebrile man with purplish plaques. Our impression was that he had vasculitis and not warfarin necrosis, which had been suspected by the cardiovascular team. The burning sensation noted by the patient lent credence to our vasculitic diagnosis. Proteinuria and hematuria were present; however, the values for blood urea nitrogen, creatinine, and glomerular filtration rate all remained within reference range. His signs and symptoms responded dramatically to prednisone. He remains on 1 mg of prednisone daily and a nephrologist continues to monitor renal function as an outpatient.

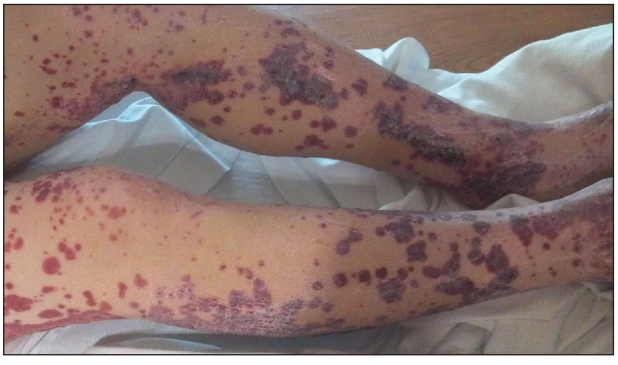

Figure 1. Henoch-Schönlein purpura. Numerous palpable purplish plaques on the bilateral legs.

Dr. J.A. Gross is from St. George's University School of Medicine, Grenada, West Indies. Dr. Grady is from DeBusk College of Osteopathic Medicine, Lincoln Memorial University, Harrogate, Tennessee. Dr. Sanchez is from Dermpath Diagnostics, Pompano Beach, Florida. Dr. D.J. Gross is in private practice, St. Augustine, Florida.

The authors report no conflict of interest.

Correspondence: David J. Gross, MD, 1100-3B, S Ponce De Leon Blvd, Saint Augustine, FL 32084 (dmjn@comcast.net).

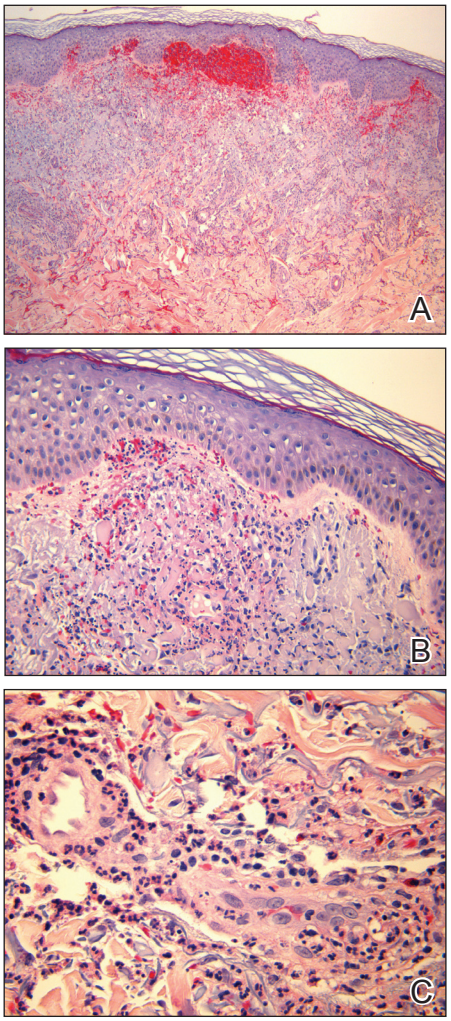

Figure 2. Henoch-Schönlein purpura. Acute neutrophil-rich perivascular and interstitial inflammation with vascular disruption of superficial vascular plexus and red blood cell extravasation (A)(H&E, original magnification ×50). Early leukocytoclastic vasculitis of a papillary dermal vessel (B)(H&E, original magnification ×200). High magnification of a superficial vascular plexus with leukocytoclastic vasculitis with fibrinoid necrosis of the vessel wall (C)(H&E, original magnification ×400).

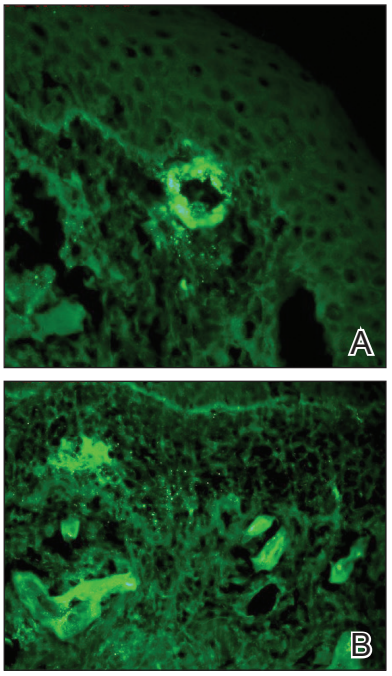

Figure 3. Henoch-Schönlein purpura. Direct immunofluorescence of IgA deposition in a papillary dermal vessel (A)(original magnification ×400) and a superficial dermal vascular plexus with IgA deposition in vessel walls (B)(original magnification ×400).

Henoch-Schönlein purpura is a systemic leukocytoclastic vasculitis involving small vessels. The small vessel vasculitis is associated with IgA antigen-antibody complex deposition in areas throughout the body. Palpable purpura typically is seen on the skin, which characteristically involves dependent areas such as the legs and the buttocks. Lesions normally are present bilaterally in a symmetric distribution. Initially, the lesions develop as erythematous macules that progress to purple, nonblanching, palpable, and purpuric plaques.[1] Henoch-Schönlein purpura most commonly involves the skin; however, other locations for the immune complexes include the gastrointestinal tract, joints, and kidneys.[2] The cause for the body's immunogenic deposition response is unknown in a majority of cases.

Henoch-Schönlein purpura most commonly is seen in the pediatric population with a predilection for males.[3] The incidence in the pediatric population is 13.5 to 20 per 100,000 children per year; HSP is more rare in adults.[4-6] Henoch-Schönlein purpura most often is a self-limiting disease that requires only supportive treatment. The signs and symptoms last 4 to 6 weeks in most patients and resolve completely in 94% of children and 89% of adults.[7] Renal involvement carries a worse prognosis. Adult patients have a higher incidence of renal involvement, renal insufficiency, and subsequent progression to end-stage renal disease.[3,8-10] In a study by Hung et al[8] of 65 children and 22 adult HSP patients, 12 adults presented with renal involvement in which hematuria or proteinuria were present. Of them, 6 progressed to renal insufficiency (defined as having a plasma creatinine concentration >1.2 mg/dL).[8] Fogazzi et al[11] reported similar findings; 8 of 16 patients affected with HSP progressed to renal insufficiency with creatinine clearances ranging from 31 to 60 mL/min, and 3 patients required chronic dialysis. Pillebout et al[9] evaluated 250 adults

with HSP and 32% reached renal insufficiency with creatinine clearances of less than 50 mL/min, with 11% of patients developing end-stage renal disease. The degree of hematuria and/or proteinuria has been shown to be an effective prognostic indicator.[9,10] Coppo et al[10] found a similar prognosis among children and adults with HSP-related nephritis.

Our patient described the burning sensation as occurring bilaterally from the knees down to the feet, which provided an additional clue that small vessel vasculitis was involved, as occluded blood vessels can cause ischemia to nerves and perivascular involvement can affect nearby neural structures. Sais et al[12] demonstrated that paresthesia in the setting of HSP was a risk factor for systemic involvement. Of note, our patient's paresthesia lasted only several days.

The cause of HSP is not always as evident in the adult population as in the pediatric population. Early diagnosis of HSP in adults may allow for the proper instatement of treatment to deter long-term renal complications. Follow-up with urinalysis is recommended because a small percentage of patients have a late progression to renal failure.[13,14]

Because the dermatologists involved in this case knew where and what types of biopsies to perform, a correct diagnosis was obtained quickly, allowing for the correct therapeutic intervention. After the diagnosis of HSP is made in an adult, nephrology should be consulted early in the treatment course.

REFERENCES

1. Rai A, Nast C, Adler S. Henoch-Schönlein purpura nephritis. *J Am Soc Nephrol*. 1999;10:2637-2644.
2. Helander SD, De Castro FR, Gibson LE. Henoch-Schönlein purpura: clinicopathologic correlation of cutaneous vascular IgA deposits and the relationship to leukocytoclastic vasculitis. *Acta Derm Venereol*. 1995;75:125-129.
3. Garcia-Porrua C, Calvino MC, Llorca J, et al. Henoch-Schönlein purpura in children and adults: clinical differences in a defined population. *Semin Arthritis Rheum*. 2002;32:149-156.
4. Stewart M, Savage JM, Bell B, et al. Long term renal prognosis of Henoch-Schönlein purpura in an unselected childhood population. *Eur J Pediatr*. 1988;147:113-115.
5. Watts RA, Scott DG. Epidemiology of the vasculitides. *Semin Respir Crit Care*. 2004;25:455-464.
6. Gardner-Medwin JM, Dolezalova P, Cummins C, et al. Incidence of Henoch-Schönlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. *Lancet*. 2002;360:1197-1202.
7. Blanco R, Martínez-Taboada VM, Rodríguez-Valverde V, et al. Henoch-Schönlein purpura in adulthood and childhood: two different expressions of the same syndrome. *Arthritis Rheum*. 1997;40:859-864.
8. Hung SP, Yang YH, Lin YT, et al. Clinical manifestations and outcomes of Henoch-Schönlein purpura: comparison between adults and children. *Pediatr Neonatol*. 2009;50:162-168.
9. Pillebout E, Thervet E, Hill G, et al. Henoch-Schönlein purpura in adults: outcomes and prognostic factors. *J Am Soc Nephrol*. 2002;13:1271-1278.
10. Coppo R, Mazzucco G, Cagnoli L, et al. Long-term prognosis of Henoch-Schönlein nephritis in adults and children. Italian Group of Renal Immunopathology collaborative study on Henoch-Schönlein purpura. *Nephrol Dial Transplant*. 1997;12:2277-2283.
11. Fogazzi GB, Pasquali S, Moriggi M, et al. Long-term outcome of Schönlein-Henoch nephritis in the adult. *Clin Nephrol*. 1989;31:60-66.
12. Sais G, Vidaller A, Jucgla A. Prognostic factors in leukocytoclastic vasculitis. a clinicopathologic study of 160 patients. *Arch Dermatol*. 1998;134:309-315.
13. Kraft DM, McKee D, Scott C. Henoch-Schönlein purpura: a review. *Am Fam Physician*. 1998;58:405-408.
14. Narchi H. Risk of long-term renal impairment and duration of follow up recommended for Henoch-Schönlein purpura with normal or minimal urinary findings: a systematic review. *Arch Dis Child*. 2005;90:916-920.